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Association of Prune Belly With Cleft Lip

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INTRODUCTION

The term "prune belly" was coined for the shriveled prune-like appearance of the abdominal wall owing to the deficiency in the abdominal musculature [Nakayama et al., 1984; Greskovich and Nyberg, 1988]. We report a patient with prune belly and cleft lip. This is the first report of this association.

CLINICAL REPORT

The patient was a female infant, the third child of a healthy 34-year-old mother and 35-year-old father. The pregnancy was uncomplicated, and the mother had no history of medication or drug use. There was no family history of cleft lip or other congenital abnormalities.

The child was born by secondary cesarean section at 38 weeks of gestation, and weighed 3,265 g. She was found to be in severe respiratory distress (Apgar scores: 1 and 3), and required intubation. In the Intensive Care Nursery, very lax and redundant skin was noted on the abdominal wall, and intestinal peristalsis could be seen clearly. She also had pulmonary hypoplasia, cleft lip, and megaloclitoris (Fig. 1). Computed tomography and ultrasonography of the abdomen showed normal size of kidneys. There was no ascites or renal dysfunction. Chromosomes were normal (46,XX).

At 10 months, the lip was repaired because of a slight improvement in the respiratory status and her parents' request. The early postoperative period was uneventful, but the respiratory function deteriorated slowly. She died at 14 months.

DISCUSSION

Abdominal muscle deficiency has been reported to be commonly associated with urinary tract abnormalities and cryptorchidism. This triad is known as prune belly syndrome (PBS). Numerous cases of abdominal muscle deficiency have been reported using the nomenclature of PBS. Most patients reported to have PBS have had the triad. However, there are some reported cases in which there were no associated urinary tract abnormalities or cryptorchidism. Such patients with an incomplete triad have recently been said to have pseudoprune [Duckett, 1986] or prune belly phenotype [Greskovich and Nyberg, 1988], and should simply be diagnosed as prune belly (PB) [Nakayama et al., 1984]. On this basis, the patient we treated would have PB associated with cleft lip. Reported anomalies in PBS are mainly gastrointestinal, skeletal, and cardiovascular malformations, and pulmonary hypoplasia [Wigger and

Fig. 1. Neonate with cleft lip, prune belly, megaloclitoris, and pulmonary hypoplasia.

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Blanc, 1977]. Not a single case of cleft lip has previously been reported in association with PBS or PB.

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